THE MECHANISM OF HAEMOPTYSIS IN MITRAL STENOSIS

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Haemoptysis is a fairly frequent event in the course of mitral stenosis, occurring in 10-20% of cases. 'Serious' pulmonary haemorrhage occurs in about 3 cases per 1,000.[4] Usually the quantity of blood coughed up is small and the haemorrhage evanescent, but as much as a pint may be voided during one episode.[3] Death from massive haemoptysis is uncommon, and when it occurs it is usually due to suffocation. The haemoptysis by itself does not materially influence prognosis except possibly in those cases evidencing marked cardiac enlargement and congestive failure.[6]

The commoner causes of pulmonary haemorrhage in mitral stenosis are (1) pulmonary oedema, (2) pulmonary infarction, (3) primary pneumonia, (4) acute cardiac failure, and (5) paroxysmal pulmonary haemorrhage. It is with the last that this paper is concerned.

PAROXYSMAL PULMONARY HAEMORRHAGE

The aetiology of paroxysmal pulmonary haemorrhage is obscure and it is believed that the case reported hereunder may help to dispel some of the existing confusion. The response to therapy although possibly fortuitous, is interesting. Theories on the mechanism of haemoptysis in paroxysmal pulmonary haemorrhage vary according to the suspected site of bleeding:

1. *Diapedesis of red cells with intact capillaries.* Presumably this is associated with high pressure in the pulmonary circuit, since it is known that changes in left auricular pressure are accurately reflected in the pulmonary capillaries.[3] If this theory is correct then the sputum of these patients should show persistent staining, and this is not so.

2. *Great rises in pulmonary capillary pressure, resulting in rupture of capillaries and consequent haemorrhage.* McGinn and White[5] studied 10 cases of acute pulmonary congestion in pure mitral stenosis. Of these, 5 had haemoptysis and 5 wheezing attacks only. Of all the attacks suffered by these patients, 70% were preceded by paroxysmal tachycardia and 80% by exertion (with tachycardia). McGinn and White therefore concluded that the conditions produced by a strong right ventricle, together with tachycardia and a stenosed mitral orifice resulted in pulmonary oedema or frank haemoptysis. These conditions would produce a rise in pulmonary capillary pressure, with possible rupture and haemorrhage (depending on local capillary fragility). On the other hand, Wolff and Levine,[7] in a study of 50 cases, concluded that 'predisposing factors were conspicuously absent'. Of their series, 46% had pulmonary infarction, which accounted for the haemorrhage. It is to be noted that 44% showed chronic auricular fibrillation, and 6% paroxysmal auricular fibrillation; and one patient had a haemorrhage during a bout of paroxysmal auricular tachycardia. Thus a good percentage of their patients were probably subject to rapid heart rates at various times, apart from any exertion. Wolff and Levine concluded that the haemoptysis was due to pulmonary hypertension with rupture of a 'sclerotic vessel'. (They admitted that the possibility of rheumatic infection of the pulmonary vessels was not investigated despite the fact that there was known to be active rheumatism in 24% of the cases). They thought that tachycardia was important as the right ventricle puts out more blood than can be passed through the stenosed mitral orifice.

3. *Rheumatic activity in small arteries, capillaries and veins, with necrotizing lesions and consequent weakening and rupture of the walls.* Little definite seems to be known about this aspect of the mechanism of haemoptysis. Brenner[1] says that the capillaries and vessels are sometimes involved in a necrotic process and rupture easily, causing early haemoptysis in cases of rheumatic carditis. Small arteries are said to be involved in the rheumatic process in 20-40% of cases of active rheumatism.

4. *Rupture of an arteriosclerotic pulmonary vessel, with or without aneurysm formation.* The arguments against this as a common cause are similar to those in the next paragraph on varices.

5. *Rupture of bronchial varices.* Ferguson, Kobilak and Deitrick[3] injected the pulmonary veins with particulate matter too coarse to enter the capillaries, hoping to demonstrate communicating channels between the pulmonary veins and the bronchial veins. In normal controls anastomoses were clearly demonstrated as small spider-like venules in the submucosa, well away from the lumen. In the cartilage region the channels were somewhat larger. No dye entered the pulmonary arteries or capillaries, nor the bronchial arteries or capillaries. Non-rheumatic cardiac cases (hypertensives, syphilitics) showed larger bronchial veins than the controls. Six of 11 cases of mitral stenosis showed 'definite evidence of enlarged bronchial veins', 4 showed greatly dilated bronchial veins along the longitudinal axis of the large bronchi. Normally blood flow is thought to be from the bronchial veins to the pulmonary veins. In mitral stenosis (the authors believe), the direction of flow is reversed, the blood passing through the azygos,

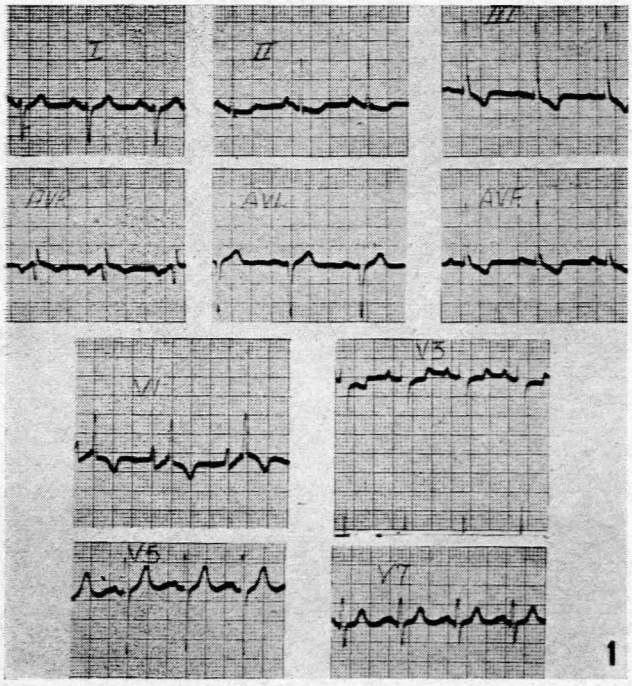
hemiazygos and intercostal veins to the right heart. Dilatation of the anastomotic channels ensues. Ferguson *et al.* conclude that 'the haemoptysis which often accompanies mitral stenosis, yet is not associated with pulmonary oedema or infarction, is probably caused by rupture or ulceration of these enlarged veins. This haemoptysis resembles massive bleeding from haemorrhoids and oesophageal varices, which, like the bronchial veins, are submucous shunts between large venous drainage areas. A severe coughing spell, mild ulcerative bronchitis or a rise in left auricular pressure could initiate such attacks of haemoptysis.' It is significant that of the present authors' 4 cases, the one surviving the longest (survival times: 1 day, 4 weeks, 3 months and 9 years) had the most marked evidence of collateral bronchial venous flow. This indicates that any collateral channel established would rather act to protect the pulmonary capillaries from acute rises in left auricular pressure, which are normally mirrored exactly in the capillaries.[2]

In a series of 168 cases of haemoptysis described by Thompson and Stewart[6] 'several cases' were bronchoscoped but no varices were seen. When the fatal case of Isaacs *et al.*[4] was submitted to autopsy, no bleeding points were found, but these workers describe chronic passive congestion in the lungs and 'marked intra-alveolar haemorrhages'. No definite vascular dilatations were found and the alveoli were filled with extravasated blood.

CASE REPORT

The patient, aged 31, a checker on the Railways, was first seen by one of us (G.R. McL.) in May 1950. At that time he presented with symptoms of angina of effort, precipitated by running, and relieved by rest. He complained of postural dizziness and was accustomed to sleep on two pillows. Physical examination revealed the following salient features: marked pallor, venous pressure slightly raised, blood pressure 85/55 mm. Hg. There was no oedema and the liver was not felt. The apex-beat was in the 5th left interspace 1¼ inches outside the mid-clavicular line. The murmur of mitral stenosis was present and an aortic leak was regarded as a possibility. ECG showed marked auricular hypertrophy. Hb 15·2 g.%. Red blood count 5·16 million per c. mm., leucocytes 12,200 per c. mm. and Wassermann reaction negative. On 1 February 1952 the liver was found to be enlarged. A 'battery' of liver function tests gave normal results. ESR 18 mm. per hour (Wintrobe), and Hb 14 g.%.

The patient was again seen by one of us (R.M.) on 28 April 1953, when he gave a history of haemoptysis, occurring roughly once a month for the past 2 years, and often brought on by running. He maintained that he was dyspnoeic on effort, but that there had been no progression of this symptom. He had no swelling of the feet; he slept on 3 or 4 pillows because he had been told to do so by his doctor, but was quite comfortable when sleeping without any. He had occasional palpitation but denied any anginal symptoms. He complained of pain over the liver area. On examination the venous pressure was found not to be raised and the lung fields were clear. The liver edge was felt ½ inch below the costal margin, and was tender. There was a rocking of the chest with systole, and the apex beat had a tapping quality. There was also a marked palpable impulse under the xiphoid cartilage.



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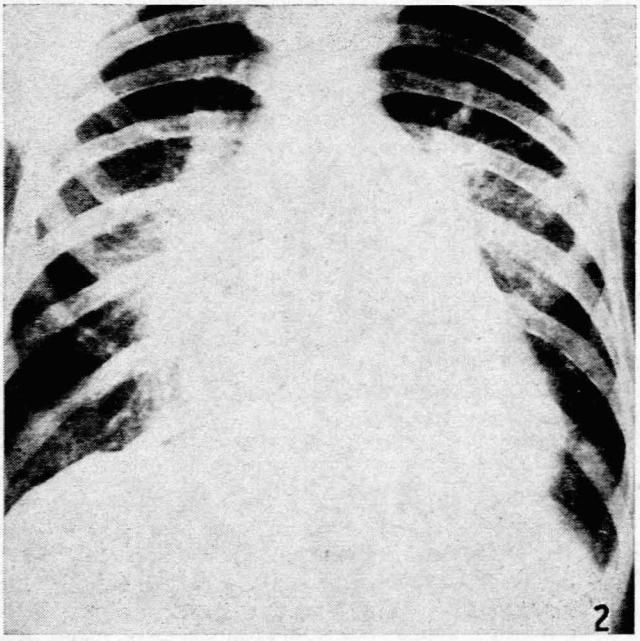
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Ausculation revealed a loud, high-pitched murmur occupying the whole of systole, best heard in the axilla. A low-pitched rumbling diastolic murmur was noticed to come and go. The blood pressure was 110/80 mm. Hg, and the ECG (Fig. 1) showed evidence of marked auricular and right ventricular hypertrophy. The chest skiagram is reproduced in Fig. 2. Screening of the heart showed a marked systolic expansion of the left auricle. Laboratory investigations showed the following:

(1) Relative plasma viscosity 1·93, normal control 1·70, water constant 1·00.

(2) Hb 14·6 g.%; erythrocytes 4·99 millions and leucocytes 7,200 per c. mm. (neutrophils 70%); P.C.V. 47%; E.S.R. 24 mm. in 1 hour (Wintrobe). (3) Gamma globulin 1·21 g.%. (4) The serum contained 25 units per c.c. of streptococcal antihaemolysin 0.

The patient was given no treatment and was not seen again until 10 June 1953. On that day at 6 p.m. he suddenly began to cough up large amounts of blood-stained froth and pure blood. His doctor admitted him to a nursing home at 11 p.m., when he was seen by one of us (R.M.). He was distressed and dyspnoeic and was coughing persistently, producing mainly pure blood but occasionally bloody froth. Temperature 100°F. Crepitations throughout both lung fields. Ausculation of the heart revealed

only a tachycardia of about 120 beats per minute and a systolic murmur referred to above; no diastolic murmurs were heard. The calf muscles of the right leg were tenser than those of the left. and Homans' sign was positive on the right. No localized tender areas could be palpated and there was no tenderness in the groin or along the course of the veins.

The clinical diagnosis was that of a ruptured atheromatous pulmonary vessel (? aneurysm) with massive haemorrhage. Pulmonary embolism was considered as a possibility, especially in view of the signs present in the right leg, but it was felt that even if this were the case anticoagulants were contra-indicated by the profuseness of the bleeding. The patient was given ¼ gr. of morphine and oxygen was administered.

At 5.30 a.m. on 17 June a transfusion of 500 c.c. of whole blood was given and this was followed by 4 pints (2·2 litres) over the next 12 hours, because of exsanguination. No improvement was noted and as the patient's dyspnoea became extreme, he was placed in an oxygen tent. Throughout the morning the haemoptysis continued unabated and at midday a pneumoperitoneum of 2,000 c.c. of air was induced by one of us (A.I.L.) in the hope of arresting the bleeding. This resulted in a marked increase in the patient's dyspnoea with abdominal pain and no decrease in the haemoptysis. At 4.30 p.m. the pneumoperitoneum was discontinued and 1,800 c.c. of air was removed. The haemoptysis continued unabated, and it was therefore decided that bronchoscopy should be done in the hope of finding an isolated bleeding point that could be dealt with.

The bronchoscopy was carried out under local anaesthesia at 6 p.m. by one of us (A.I.L.) and findings were as follows: 'A large quantity of blood was aspirated from the bronchial tree on both sides. After aspiration blood continued to reaccumulate on both sides from the upper as well as the lower lobe orifices. The bleeding was obviously bilateral and generalized and did not originate from any single source. After bronchoscopy the throat rattle disappeared and the dyspnoea was relieved'. It must be noted here that nothing was done during the bronchoscopy which could possibly have influenced the bleeding.

At 8.30 p.m. the bleeding was as strong as ever and the patient's general condition poor. It was felt that the bleeding was probably the result of a general capillary oozing and so he was given 150 mg. of cortisone by mouth, and a further 150 mg. at 10 p.m. He was also given 1 g. of ascorbic acid by injection and 60 mg. of Rutin 4-hourly. At 10 p.m. (i.e. 1½ hours after the first dose of cortisone), the bleeding ceased, and it did not recur. Small amounts of dark altered blood were coughed up during the next 2 days. Over the next few days all drugs were gradually tailed off and then stopped, and the patient was discharged on 20 June.

In view of the tendency towards massive haemoptysis and the evidence of marked pulmonary hypertension, and also the now persistently-raised venous pressure, it was decided to perform a mitral valvotomy. This was done on 17 July 1953 by one of us (A.I.L.). At operation a regurgitant stream was felt and the mitral valve was found to be tightly stenosed and calcified. It admitted only the tip of the index finger. An adequate split was obtained. At no time after the operation was the patient's condition satisfactory and he died on the 3rd post-operative day in a state of peripheral circulatory failure. Permission for a post-mortem examination was not obtained.

CONCLUSION

The following facts in the above case are important, since they point towards a definite mechanism for the haemoptysis:

1. Tachycardia as produced by effort, was regularly associated with bouts of haemoptysis.

2. During the patient's massive haemoptysis (5 pints) tachycardia was again present.

3. From the onset of bleeding the patient had remained in a sitting position, or at most inclined backwards some 10-20 degrees, supported by pillows. Yet at bronchoscopy blood was seen pouring from *all* the bronchial orifices in both lungs, from the upper as strongly as from the lower.

4. The response to cortisone was dramatic; within 1½ hours of the first dose, no fresh bleeding occurred.

5. At bronchoscopy no varices were seen.

6. Rheumatic activity was known to be present.

Conclusion. Analysis of the evidence available on the mechanism of haemoptysis in mitral stenosis, leads us to believe that paroxysmal haemorrhage may best be accounted for by the presence of a stenosed mitral valve, a strong right ventricle, and tachycardia. Probably—although there is no direct proof of this—rheumatic activity in the smaller pulmonary capillaries plays an important part. This set of circumstances will cause rupture of the pulmonary capillaries with intra-alveolar haemorrhage, sometimes with pulmonary oedema, and this results in the coughing up of blood. The process is a generalized one, and bleeding from a particular point is entirely conjectural. Broncho-pulmonary venous shunts undoubtedly appear to exist, but their influence is mainly protective.

The only indication for bronchoscopy is the relief of dyspnoea due to accumulation of blood in the bronchial tree.

SUMMARY

The sources of blood in the sputum in cases of mitral stenosis are enumerated.

2. The mechanisms of paroxysmal pulmonary haemorrhage are analysed and discussed.

3. A case of massive haemoptysis (5 pints) is presented, with bronchoscopic findings pointing to a definite aetiology.

4. The effect of treatment with cortisone, although possibly fortuitous, suggests that further trials are worth while.

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